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# Newborn intestinal obstruction due to mesenteric lymphangioma: A diagnostic challenge

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ABSTRACT

Lymphatic malformations are uncommon benign cystic lesions that typically present in the neck and axilla during infancy. They are more rarely observed in the abdomen and extremities. This study presents the case of a newborn infant who was admitted to the hospital with vomiting. While the cause of the vomiting was investigated, a complete intestinal obstruction developed, and an emergency surgical intervention was performed. During a laparotomy, a cystic mass originating from the mesentery was discovered. A lymphangioma mass resulting in intestinal obstruction during the third week of life was not considered in the differential diagnosis.



## 1. Introduction

Lymphangiomas are tumors of the lymphatic system and are characterized by anastomosing lymphatic channels and cystic spaces varying in size; the diameter of such a mass is usually smaller than 0.5 cm, but they can reach much larger sizes [1]. These tumors occur due to an abnormal connection between lymphatic ducts and the venous system during the fetal period [2] and can be observed in any part of the body. Tumors are usually located in the neck, with a rate of 75%, and in the axilla, with a rate of 20%. Lymphangiomas occur less often in the thoracic and intra-abdominal visceral organs, with a rate of 4–5%. They affect the mesentery, omentum and retroperitoneal space at a rate of less than 1% [3]. The clinical symptoms depend on the size and location of cysts. This report presents the case of a male newborn with mesenteric lymphangioma and an unachievable preoperative diagnosis. Cases of mesenteric lymphangioma and newborn bowel obstruction in the literature were also reviewed.

## 2. Case report

A 20-day-old male infant was brought to the hospital after one week of vomiting, being unwilling to feed and passing less stool. The full-term infant was born vaginally to a 30-year-old mother with a gravidity of three and a parity of three. His birth weight was 2500 g. No specific features were present in the family history, and prenatal follow-up included ultrasonography (US). The infant did not present any complaints during the first two post-natal weeks; his feeding and defecation were normal, and the symptoms occurred and progressed in week three. On physical examination, mild dehydration, respiratory distress and abdominal distension were observed. No palpable abdominal mass was found. Mid-penile hypospadias and chordee were noted. Blood biochemistry and other laboratory results were normal. No metabolic alkalosis was observed. An erect plain abdominal radiograph demonstrated only a wide-based stomach with an air-fluid level and no gas or calcification in the remaining abdomen; the intestines were not visible on the erect radiograph (Fig. 1). US examinations were repeated twice under emergency and elective conditions; the resulting differential diagnoses included malrotation, annular pancreas or midgut volvulus. During that period, non-bilious vomiting gradually became bilious, and pyloric stenosis, an initial diagnosis, was eliminated. A contrast upper gastrointestinal (UGI) series was performed to evaluate the possibility of incomplete duodenal obstructions, such as type 1 atresia, intraluminal duodenal web (wind sock), superior mesenteric artery syndrome, intestinal malrotation-Ladd's bands, midgut volvulus, and duodenal stenosis. The contrast UGI results showed an enlarged stomach with contrast material retention, nondilated loops of the small bowel and no evidence of a double bubble (Fig. 2).

On day 5 after admission, a complete intestinal obstruction developed with a gradual deterioration in general condition. The amount of bilious gastric aspirate and abdominal distension

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**Abbreviations**

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| US | ultrasonography |
| UGI | upper gastrointestinal |
| CT | computed tomography |

increased, stool discharge ceased, and an empty rectum was detected on rectal examination. Emergent surgical exploration using a right transverse incision above the level of the umbilicus was performed. Intraabdominal serous fluid was noted, and a sample was collected. A lymphangioma approximately 8 cm in diameter was observed; it had developed in the mesentery of the 7 cm proximal jejunum segment of the small intestine and completely occluded the intestinal lumen. The lymphangioma had thin walls, contained chylous fluid, and exhibited hemorrhagic areas in various sections; in addition, dilated lymph channels were visible (Figs. 3–5). No other pathology was observed during the abdominal exploration. The multiseptated cyst was excised along with the small intestine segment that it surrounded. Intestinal continuity was maintained with end-to-end anastomosis. Although no malrotation was observed, an elective appendectomy was performed after a risk and benefit assessment (Fig. 6). An intraabdominal drain was not inserted. The nasogastric catheter remained for 48 h. Oral feeding began on the third day. The patient was discharged on day 7. Growth culture and fluid cytology results were negative for the fluid from the abdominal cavity. A gross examination of the specimen showed enlarged lymph vessels, which contained lymph fluid. The diagnosis of lymphangioma was confirmed by a histopathological analysis. The cystic mass, which was completely located within the mesenteric boundaries, had a type 2 Losanoff pathological classification. The patient remained asymptomatic during a follow-up period of three years.


**Fig. 1.** Erect plain abdominal film showing a gasless abdomen with the exception of the air-fluid level in the stomach.

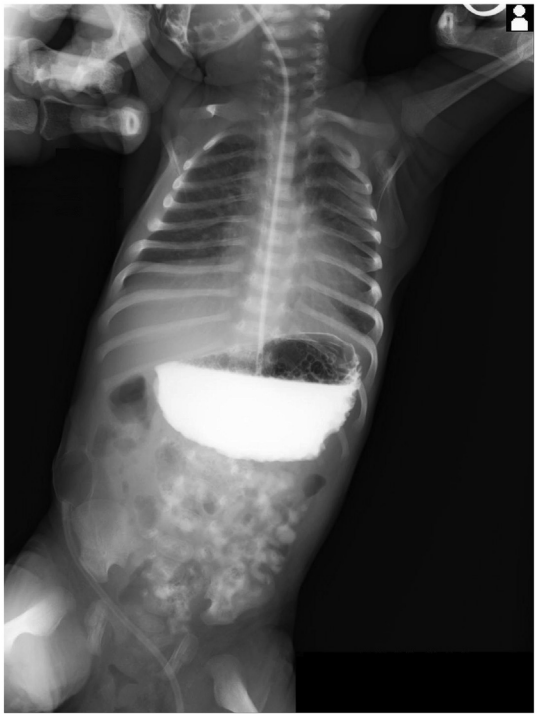

**Fig. 2.** Contrast UGI results showing a dilated stomach and a normal bowel configuration.

## 3. Discussion

The incidence of lymphangiomas is not precisely known because some cases are asymptomatic. The patient presented in this study was male, and hypospadias existed as an additional abnormality. The incidence of this condition in males and females varies according to reports in the literature. In this work, an electronic search of the English literature was performed to screen for relevant papers and calculate the male:female ratio; the 8 largest studies considering only mesenteric lymphangiomas included a total of 110 patients, and the male:female patient ratio was calculated to be 1.5/1 (66/44) [2,4–10]. Few associated pathologies were found in the search, including vascular malformation in three studies [4], Russell-Silver syndrome in one study [7], left renal agenesis in one study, pancreatic and renal cysts in one study [10], and intestinal malrotation in four studies [11–13]. No additional abnormalities have been reported in other case reports or series. In contrast to Antoniou et al., Weeda et al. suggested in a report of 2 cases that mesenteric lymphangiomas could be an acquired pathology that occurs as a result of malrotation and volvulus [11–13]. In this case, the cecum was in its normal location, and no malrotation of the intestine was present. Obstruction occurred due to extrinsic pressure instead of volvulus. This study supports the idea that mesenteric lymphangioma is an isolated disease; although it is a congenital disease, it is observed in generally healthy individuals [14], and the clinical symptoms can occur at any time.

Of all lymphangiomas, 65% occur at birth, and 90% occur during the first two years of life [3]. They can remain asymptomatic for a lifetime or can produce life-threatening complications. While small intraabdominal lymphangiomas cause only the accumulation of a small volume of peritoneal fluid, symptoms will occur if the mass increases in size [1]. They are characteristically slow-growing lesions but can also reach alarming sizes due to rapid massive growth

**Fig. 3.** Intraoperative findings showing the lymphangioma mass.

**Fig. 5.** Pathological specimen of the mass, which was completely removed with the involved jejunal bowel segment.

[15]. Patients with mesenteric lymphangiomas are typically hospitalized at early ages due to acute abdominal conditions, such as intraabdominal bleeding, volvulus or intestinal obstruction, as in this case [4,16–18]. However, patients who remain undiagnosed as adults are typically hospitalized due to chronic complaints. According to a report by Aprea et al. of the oldest patient series (65–80 years) in the literature, these patients were admitted to the hospital due to chronic abdominal pain and swelling [10].

During the neonatal period, most abdominal masses are retroperitoneal and of renal origin; therefore, they are identified by the doctor or family on the first day of life [19]. In this patient, the mass was located in the intraperitoneal space, and the patient was hospitalized at 3 weeks of life with a complaint of vomiting; therefore, the mass could not be diagnosed preoperatively.

Most patients who are referred to the neonatal unit for congenital surgical reasons present with bowel obstruction. The four cardinal signs of surgical bowel obstruction are maternal polyhydramnios, bilious vomiting, delayed meconium passage and abdominal distension [20]. The present patient exhibited only abdominal distension and non-bilious vomiting during hospitalization.

Neonatal intestinal obstructions occur due to extrinsic, mural or intraluminal causes [16]. Mural pathologies related to failed canalization of the intestinal lumen are observed in approximately 95% of patients who undergo surgery due to intestinal obstruction during the neonatal period [21]. Obstruction of the intestinal lumen by an extrinsic mass such as a lymphangioma is rarely observed during this period. Verma et al. conducted a study during a 15-year period to examine 298 patients who underwent surgery due to neonatal intestinal obstruction, and they reported that a cystic lymphangioma resulted in intestinal obstruction in only 1 neonatal patient [22]. Obstructions that occur due to extrinsic mass pressure usually cause symptoms upon growth of the mass; the process begins with non-specific semi-obstructive symptoms, such as not ingesting milk, intermittent vomiting that transitions to vomiting with bile, and decreased defecation. However, symptoms of complete gastrointestinal obstruction occur as complete pressure obstructs the intestinal lumen upon growth of the mass.

Abdominal X-rays can reveal bowel loops displaced by a mass and/or calcifications in the cyst wall; however, X-rays alone are insufficient for evaluating vascular malformations [3,23,24]. The direct abdominal X-rays of this patient did not reveal bowel displacement and/or calcification due to the mass. Therefore, the possibility of an intestinal lymphangioma-like mass was not considered.

As lymphangiomas are slow-flow vascular malformations, they can be best identified by US [23]. Many cases of mesenteric lymphangioma diagnosed with US have been reported in the literature [25,26]. In cases of lymphangiomas, US examinations can show the macrocystic and microcystic components with good contours separated by septa, the multiloculated structure, the wall thickness

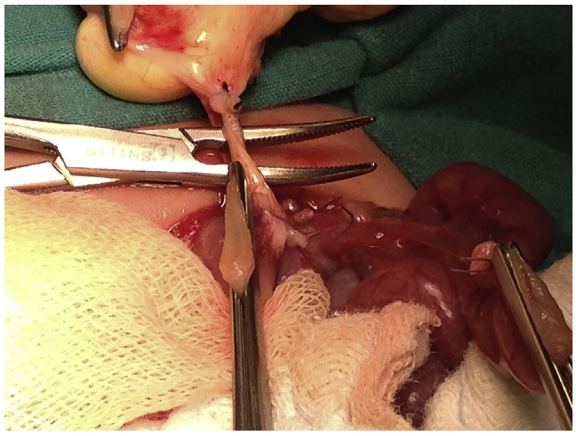

**Fig. 4.** Intraoperative findings showing the dilated lymph channels.

**Fig. 6.** Intraoperative findings showing the cecum located on the right inguinal region; an appendectomy and a jejuno-jejunal anastomosis were performed.

of the cyst, the existence of anechoic fluid, and the presence of ascites in the abdomen [2,3,5,23,26]. Differential diagnoses in female patients are limited due to the probability of ovarian cysts [2]. These cystic lesions, which can also be identified with antenatal US, can mimic renal cysts, enteric duplications and other intra-abdominal cysts, such as ovarian cysts, meconium ileus, meconium peritonitis, intestinal atresia and ascites [14,17,27]. Eventually a final preoperative diagnosis could not be established in our patient. Because US is a radiological examination that depends on the imaging quality of the US device, the ability of the operator and the results require interpretation. US and computed tomography (CT) have been used together in many series to confirm preoperative diagnoses [2,5,7,28]. However, a correct preoperative diagnosis could be established with CT and US in only 16 of the 25 patients in a study by Kim et al. [4]. The current patient could not be examined by CT due to technical reasons.

The last examination performed after the X-ray and US evaluations was the contrast UGI series with a water-soluble contrast agent. The X-ray series showed that the stomach was dilated, and discharge was delayed; no signs of an obstruction or a malrotated bowel configuration were visible. As the patient developed a complete obstruction at this stage, an emergency operation was performed.

Lymphangiomatous masses, which are benign tumors of the lymphatic system, do not demonstrate malignant degeneration. Vital structures should not be damaged during complete excision. Definitive treatment consists of total excision [1], and recovery is observed upon total cyst removal. Drainage inside the peritoneal cavity [1], enucleation [2] and marsupialization [6] have been suggested for cases in which total surgical removal could not be performed. Studies have also reported that asymptomatic cases that could not be surgically treated have regressed [8,14]. An increasing number of laparoscopic excisions have also been presented in recent years [8,28]. Another treatment option is sclerotherapy, in which various agents are used. Sclerotherapy can be primarily or surgically performed for cases that cannot be totally removed. Intralesionally administered doses can be repeated in sclerotherapy. The treatment period is long, and complete resolution has not been observed [14,29]. The Losanoff pathological classification is used to group mesenteric lymphangiomas into 4 types [3]. The cyst in the present patient was considered type 2, as the mass was completely removed with an involved bowel segment. It did not extend toward the retroperitoneal area. The rate of postoperative complications is very low in most mesenteric lymphangioma reports. Morbidity and mortality generally occur in cases with torsion due to intracystic bleeding [2,14,17]. Some rare postoperative complications have been observed at 1 year after surgery, including chylous ascites, adhesive ileus and recurrence [2,4,6,7]. Patients are followed up with postoperatively, both clinically and by US.

In terms of follow-up, complication-free periods of 17, 11 and 16 years have been reported in the literature [2,8,9]. In the current case, the follow-up period was 3 years.

During the antenatal period, no polyhydramnios was observed in the present patient. Following birth, his gastrointestinal passage functioned properly, and severe distension, bilious vomiting and the other complaints began during the third week. Therefore, a congenital intestinal obstruction was not considered. The initial differential diagnoses were pyloric stenosis and type 1 atresia. However, the US and X-ray results did not support these diagnoses, and the US findings did not indicate the possibility of lymphangioma. The mass gradually grew in the following days, and it caused extrinsic pressure in the proximal jejunum lumen, resulting in a complete obstruction.

## 4. Conclusion

The ratio of males to females with lymphangiomas was calculated in this study to be 1.5:1, according to studies found in a search of the literature. Mesenteric lymphangiomas usually occur in otherwise healthy individuals and are rarely associated with other congenital abnormalities. Therefore, this rare condition results in low morbidity and mortality rates.

The occurrence of intestinal obstruction in newborns who are fed normally, defecate after birth and have an open gastrointestinal passage may be due to intraabdominal lymphangiomas that do not cause any symptoms initially due to their small volumes. These masses do not have any specific signs. It is suggested that to make a correct diagnosis for similar situations in the future, better conditions in US examination and advanced imaging technologies are required. Including growing mesenteric lymphangiomas among rare causes of gastrointestinal obstructions in neonates will be helpful for early diagnosis and treatment plans.

## Conflicts of interest

The author declares that no conflicts of interest exist regarding this manuscript.

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